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FAMILY PERIODIC PARALYSIS.*

With a Report of Cases hitherto published.

By EDWARD WYLLYS TAYLOR, M.D.,

Instructor in Neuropathology, Harvard University; Assistant in Neurology, Massachusetts General Hospital; Visiting Neurologist, Long Island Hospital, Boston Harbor.

The condition which forms the subject of this communication is one of great rarity. In America it has, with two possible exceptions, remained absolutely unnoticed in the form which I shall later describe. In Europe, on the other hand, and notably in Germany, a few cases of an identical sort have been reported in detail.

The term "Family Periodic Paralysis" is evidently a clinical one, and serves merely the purpose of description. With certain modifications, this term has been generally used by the German writers, and, in the absence of definite pathological knowledge on the subject, serves its end sufficiently well.

The affection is characterized, in its typical form, by extensive, flaccid motor paralysis, associated with loss of reflex and electrical excitability, without sensory or psychic disturbance of any sort, and with intervals of perfect health. The disturbance is periodic, and the predisposition to its development is clearly hereditary. It is at once evident that such an affection has no counterpart among the known diseases of the nervous system, and should, therefore, command our warmest interest. Such interest has by no means been lacking in those who have had the fortune to observe the condition, as a study of the literature amply demonstrates.

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*Read at the twenty-fourth annual meeting of the American Neurological Association, May, 1898.

The earliest references to a disturbance analogous to that which we are considering come from Cavaré[1], 1853. and Romberg,[2] 1857. Both of these are referred to by Westphal (5) in a later article, and to his paper we owe the references, which were not obtainable at first hand. Cavaré's case occurred in a woman of 24, who had several attacks of generalized paralysis lasting from five to eight hours; the attacks were of the quotidian type, and were relieved by quinine, but no definite statement is made as to their malarial nature. Romberg describes similar attacks, in which the lower extremities were affected, and which he attributed in his case to intermittent fever. These attacks were cured by quinine.

Nearly twenty years later Hartwig (1), in his inaugural dissertation, again drew attention to the subject through the publication of a carefully observed case of what he called intermittent spinal paralysis. The patient was 23 years old. He had had malaria five years before the present attack, which came on with weakness of the legs, the arms and neck muscles being later involved, somewhat after the manner of a typical so-called Landry's paralysis, only much more rapidly. Speech, swallowing and breathing were hindered. The facial nerve was free;

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[1] Cavaré: Gaz. des Hôpit., 1853, No. 89 (aus der Gazett méd. de Toulouse), quoted by Erb. Handbuch d. Krankheiten des Nervensystems, I., p. 822, als Fall von Macario.

[2] Romberg: Lehrbuch der Nervenkrankheiten, 3. Aufl., 1857, p. 752.

sphincters uninvolved; sensibility and mental state unimpaired. Electrical reactions were almost entirely lost during the attack. Recovery took place in 24 hours. Similar attacks followed, and this patient was also much helped, but not cured, by quinine. He was observed for six months, and the supposition of a malarial cause was entertained. This case, further details of which are given in the summary later, is, in its general features, a type of the condition we are about to study.

Samuelson (2), in 1876, reported a case of intermittent paraplegia, with no disturbance of sensation, in a person of thirty-eight, in which a suspicion of hysteria was aroused, though probably not justified by the facts.

In 1882, Schachnowitsch (3) described a like condition in a patient whose father was similarly affected, and who died from increasing attacks in his fifty-fourth year. The hereditary character of the affection first appears in this observation. In the light of others' experience, death from this cause must be looked upon as unique; this would seem to throw a certain doubt upon the accuracy of the diagnosis, in spite of the fact that in Goldflam's opinion the condition described is accurately classified.

In the same year a paper was written by V. P.Gibney (4), on an unusual form of paralysis, which he regarded as distinctly of malarial origin. He speaks of two cases, both in children, in which, after a malarial attack, paralysis of the extremities came on, associated with muscular atrophy, with certain sensory and constitutional disturbances, followed by slow recovery. The late Dr. Seguin diagnosticated one at least of the cases as myelitis of the anterior horns. Gibney regarded them as similar to the conditions reported by Cavaré, Romberg and Hartwig, to which allusion has already been made. In any event, they are so clearly unlike the typical condition, to which we wish to draw attention, that in our opinion they almost certainly do not belong in the same category, whether malarial or not. They are alluded to in this connection because Westphal, in a subsequent communication, following his earlier paper, is inclined to regard Gibney's first case as belonging to the group of the family periodic paralyses. Westphal further thinks that neither of Gibney's cases was due to malaria.

The first really important consideration of the disease, if we may so call it, we owe to Westphal (5), who, in 1885, described in detail a typical case under the heading "Ueber einen merkwürdigen Fall von periodischer Lähmung aller vier Extremitäten mit gleichzeitigem Erlöschen der electrischen Erregbarkeit, während der Lähmung." In the study of this case Westphal was assisted by Oppenheim, to whom we owe much careful work on the electrical changes. The history, in outline, is as follows: The patient was a boy of twelve, with no hereditary predisposition, so far as learned. The attacks came on by weakness of the legs; three hours later the boy could not stand, and the attacks were followed in a short time by a complete paralysis of arms and legs, of a flaccid character. Knee reflexes at times were not obtainable. Cranial nerves were free. Sensation was unaffected; mind clear. Electrical examination showed distinct quantitative diminution to both galvanism and faradism, and complete loss in certain nerve distributions. Gradual improvement occurred the following day, which soon became complete, with a slow return of electrical excitability. He was perfectly normal in all respects between the attacks. Repeated attacks of this character occurred, which were carefully studied until the patient passed from observation. The condition above briefly described excited Westphal's profoundest interest. His astonishment was chiefly aroused by the extraordinary electrical and reflex changes, which at once placed the disturbance without the pale of the so-called functional diseases, notably hysteria. Westphal's since much-quoted words on the subect are: "Dieses relativ schnelle Erlöschen und Wiederkehren der electrischen Reizbarkeit in Nerven und Muskeln steht ganz einzig in seiner Art da; wir kennen weder eine Krankheit des Rückenmarks noch der spinalen Nerven, in welcher jemals etwas Aehnliches beobachtet wäre; ebenso lässt uns die Physiologie in Betreff einer Erklärung vollständig im Stich * * * Wir stehen somit dem geschilderten Krankheitsfalle als einem Räthsel gegenüber, und sind nicht einmal im Stande, eine annehmbare Hypothese aufzustellen, weder über die Natur der in grösseren Intervallen (nicht nach dem Wechselfieber Typus) auftretenden Lähmungserscheinungen, geschweige denn über die Ursachen des schnellen Erlöschens und der ebenso schnellen Wiederkehr der electrischen Erregbarkeit der Nerven und Muskeln."[3]

Following Westphal, in the same year, Fischl (6) adds another interesting observation, which, although differing in certain respects from other reported cases, probably belongs to the same general group. The patient was a girl of eight, also without hereditary history of significance. There was a complete flaccid paralysis of the legs, with loss of knee-jerk, the arms being very slightly, if at all, affected; recovery in twenty-four hours. At a subsequent attack, faradic examination showed a complete loss of reaction in muscles and nerves involved. Condition between the attacks perfectly normal. Certain peculiarities of this case are of special interest. These will be hereafter discussed.

In the following year, 1886, Westphal (7) published a supplementary article, in which he discussed Gibney's paper, which he had before overlooked. Beyond this, the communication adds nothing to our knowledge.

Cousot (8, 9), in 1886, and again in 1887,[4] gives histories of a number of cases occurring in one family. The mother and four children were affected; beyond this the heredity does not go. The mother often lost her motor power once a week, an affliction which lasted through life.

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[3] Westphal: Ref. 5, p. 511.

[4] We quote only from the latter article. Ref. 9, of bibliography.

The children suffered from similar attacks, motor weakness to point of paralysis, arms and legs both affected, loss of electrical reactions; sensation and intelligence undisturbed; normal between attacks.

Griedenberg (10) adds another case to the list, that of a man of twenty-two, who had periodic attacks of an identical sort with those already described.

In 1890 Goldflam (11) first published his investigations on the subject with the title, "Ueber eine eigenthümliche Form von periodischer, familiärer, wahrscheinlich autointoxicatorischer Paralyse." With the attention which Westphal had already brought to the matter, this and the later work of Goldflam and Oppenheim afford us what knowledge we have of this unique affection. To Goldflam, however, in particular, belongs the credit of working out in minutest detail the clinical phenomena, and also of instituting various experimental researches in the hope of elucidating the subtle problems of etiology. This first complete description of the disease covers eleven cases in the patient's family, and affords, therefore, an opportunity of noting the remarkable hereditary character of the disturbance. The attacks in the best observed of these cases occurred in a boy of 17, and conform in all essential regards with the cases already briefly described. The following year, 1891, Goldflam (13) further elaborated his paper in a detailed piece of work under the same title. This second article covers all the ground of the previous one, and adds considerably to it. These papers, particularly the second, are monographic in their completeness.

Between the publication of Goldflam's two papers Pulawski (12) reported a perfectly typical case, so like others that the same general description serves for all.

In the same year that Goldflam's later communication appeared Oppenheim (14) had an opportunity to observe again the case described by Westphal in 1885, and to make a much more careful examination than had hitherto been done. The chief value of this work lies in the investigation of the electrical changes, under varying conditions and in different stages of the attacks, as well as in the free intervals. His conclusions in general are that at the height of the attack nerve-muscle excitability is completely lost in certain groups, but that electrical stimulation does not always give uniform results. Oppenheim also draws attention for the first time to a hardly less remarkable phenomenon, viz., a temporary dilatation of the heart during the attack, and signs indicative of mitral insufficiency, which gave place to perfectly normal conditions between the attacks.

An interesting but cursorily reported case appeared in the University Medical Magazine for 1892-'93, observed by Burr (15). The patient was a strong man of thirty, who since his tenth year had had periodic attacks, occurring about four times yearly, and lasting from a day to a week. The long duration of the attacks, and the fact that but one half of the body was affected when seen by Burr render the case noteworthy. No electrical examination was made.

In 1894 two papers were published, by Hirsch (16) and Rich (17), of Ogden City, Utah. The mother of Hirsch's patient, who was a man of twenty-six, had similar attacks; otherwise no hereditary predisposition. Hirsch verified Oppenheim's observation of temporary insufficiency of the mitral valve during the attacks. In other respects, the case is only noteworthy from its similarity to others.

Rich's (17) communication is an exceedingly interesting one, and the subject is well deserving of more study than he has devoted to it in his published article. He describes, without citing individual cases, a peculiarity existing in his own family for five generations, of a temporary motor paralysis due to cold, affecting by preference the facial muscles, without accompanying disturbance of the mind or of sensation. An unusual exposure to cold or dampness brings on almost immediately a more or less complete disturbance of motion. He speaks of one occasion in which a person was completely paralyzed, with the exception of the tongue, through sleeping in moist underclothing. A paralysis of the tongue could easily be brought about by holding snow in the mouth, taste being unaffected. Complete recovery always takes place, preceded by a feeling of exhaustion. The paralysis is always of the nature of a tonic spasm. No mention of temporary electrical changes or of a detailed physical examination is made in his paper. Apart from the peculiarity mentioned, there was absolutely no neurotic tendency in the family, but this one idiosyncrasy had never skipped a generation, as far back as could be traced.

The analogy to the condition we are considering is so striking, and the exciting cause so definite and yet so different from other reported cases, that this paper of Rich must be regarded as most significant and important, in spite of its brevity.

In 1895, Goldflam (18) carried his researches still further, and attempted by various means to arrive at definite conclusions of an etiologic sort, with what success we shall have occasion to see later. In a footnote to this elaborate paper, he alludes to two other cases, in addition to those, eleven in number, which he had previously described, both occurring in children, and he thinks of the same character. He does not go beyond a very general statement, pending further observation. It may also be said, in passing, that Goldflam regards a case, alluded to in Erb's monograph on Thomsen's disease, by Rothe[5] as belonging to the family periodic paralyses.

Bernhardt (19), in the following year, writes on two cases in a father and son, which correspond closely, in certain respects, to the clinical picture already frequently outlined. Bernhardt, however, is chiefly interested in the association of periodic paralysis, with muscular distrophy and with other forms of well recognized disease.[6]

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[5] Rothe: Statistischer Sanitätsbericht über die Königl. Preuss. Armee, u. s. w., für 1879-81. Berlin, 1882, p. 51 (ref. taken from Goldflam's paper, bibliog. 18).

Goldflam (20) contributed still a fourth paper to the subject in 1897. In it he reports eight additional cases, in a branch of the family he had already described, and also three other cases occurring in a totally different family. His conclusions from this further study are essentially confirmatory of his previous observations, though differing somewhat in detail.

From the foregoing historical sketch, which, we believe, is complete in its references, it is evident that the condition outlined is of curiously infrequent occurrence, and that when it has been observed it has aroused an interest of the profoundest sort. The German cases have been studied with extreme minuteness by Westphal (7), Oppenheim (14), Goldflam (11, 13, 18, 20) and others; in America we find but three references, one of which, that of Gibney (4), is doubtful of classification. Of the other two, Burr's (15) communication narrates a single case, which is, no doubt, of the periodic type, but reported with less completeness than its importance warrants; and, finally, the extraordinary condition described by Rich (17), as due to exposure, presents a most interesting and close analogy to the matter under discussion. We have, therefore, no excuse to offer for the presentation of the following family history, and the somewhat detailed consideration of the affection as it occurred in two of its members:[7]

The affection in this family first developed five generations back. There is a tradition that the person afflicted was running violently, when something seemed to give way in his back, and he fell to the ground, prostrated. From that time on he was subject to attacks of a periodic character. Of his three sons, only one seems to have had the disease. The other two, with their children, were never afflicted. The son who was affected was a judge, and those who remember him cannot recall that he was ever kept from his work by the attacks, but remember that he frequently needed assistance in getting in and out of his carriage. In each succeeding generation the disturbance was transmitted to one or more members down to the present, in which two (our patients) out of three are typically affected.

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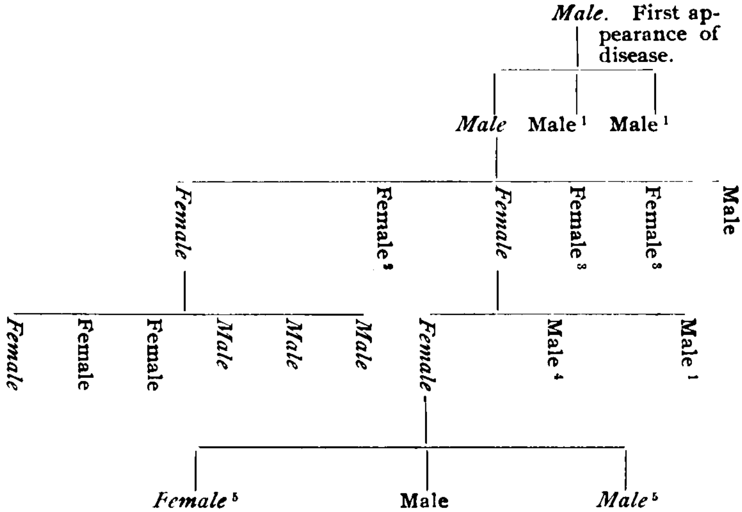
[6] See earlier paper. Bernhardt: Muskelsteifigkeit und Muskelhypertrophie (ein selbstständiger Symptomencomplex). Virchow's Archiv, 1897, lxxv., p. 516.

[7] For many of the following facts and observations I am indebted to a member of the family, a student of medicine, whose assistance has been of the greatest help in preparing the clinical side of this report.

FAMILY PERIODIC PARALYSIS.



EXPLANATION OF CHART.

Italics:	Persons affected.
Roman letters:	Persons exempt.
1.	Children exempt.
2.	Unmarried.
3.	Died young.
4.	Epilepsy.
5.	Patients studied.

The genealogy of the family is presented in the above diagram, which makes a detailed description superfluous:

The hereditary tendency is transmitted through the female as well as through the male line. Apart from this affection, there is no significant neurotic tendency observable. One person, who was exempt, had epileptic convulsions, and the mother of our patient, and, possibly, the grandmother, died of tuberculosis of the lungs. Neither of these facts is significant, inasmuch as the paralytic condition had already existed for three generations in a perfectly non-neurotic family, so far as ascertainable. It is to be remarked, further, that the family is one of rather unusual mental power.

Detailed facts regarding previous generations are difficult to obtain, although certain members of the second generation are living. The testimony of those members of the family with whom I have been able to talk is, however, definite, viz., that the attacks in all cases, with one exception, when they began at the eighteenth year, have come on at about the age of puberty, and persisted, with varying frequency, during the active years of life, after which, between the fortieth and fiftieth year, they have gradually disappeared. The character of the individual attacks has been in all cases strikingly similar, to such a degree that one description serves for all.

That description we shall give in the following most carefully observed case in the present generation:

CASE I.—J. T. S., 19; schoolboy. As a young boy he was perfectly well, and free from the ordinary diseases of childhood; no malaria nor venereal infection. During the summer of 1894, when he was 14 to 15 years old, he first noticed a certain slight feeling of weakness in the thighs and calves of the legs. His earliest remembrance is that in playing "leap-frog" he was weaker than usual. This condition continued in intermittent fashion during the summer, and in the fall of that year he had the first typical attack. It occurred at night, as all the succeeding ones, practically without exception, have done. The attack was characterized by a general weakness of the body and limbs, without further disturbance of any sort. From that time the attacks followed each other rapidly, on an average of one a week. He would awake in the morning to find himself paralyzed; he thinks he was often awakened by the paralysis. His helplessness usually continued during the day, and passed away gradually toward night. His own observation was that the attacks were brought on by excitement, exercise or late hours. Between the attacks he was absolutely well, with no trace of muscular weakness.

From that time to this, a period of nearly four years, there has been no change, with the possible exception that the attacks have increased somewhat in frequency and diminished in intensity. This is the general history of the affection, as it appeared to the patient's own observation of himself. Being an active boy, with a taste for athletic exercise, it goes without saying that he has been exceedingly annoyed by his persistent periods of disability. So many of his ancestors, including his mother, have, however, been similarly affected that he has developed a certain philosophical attitude, which helps him over days of enforced idleness.

A typical attack, in detail, is as follows: After a day usually of somewhat violent exercise, as, for example, bicycle riding, he goes to bed feeling perfectly well and strong. There are no prodromal symptoms whatever, beyond, on occasions, a certain feeling of weakness, which usually is not observed, since the attack begins almost invariably at night. The portion of the body first affected, whether legs or arms, is also not definitely known for the same reason, though he thinks a weakness of the legs is antecedent. On awaking in the morning, he finds himself in a pronounced attack, completely paralyzed, with the general exception of the cranial nerves. Voluntary movement is absolutely abolished. Coughing and sneezing are impossible, but respiration, so far as he has observed, remains normal. There is no dyspnœa. The muscular system is perfectly flaccid. He has no desire for food or drink during the attack, and he often goes twenty-four hours or more without urination. There is sometimes apparent temporary urinary retention, which he thinks is due rather to the awkward position in which he happens to be lying than to any actual disturbance with the sphincters. He never defecates during an attack. He has no sensory disturbance whatever, and his mind is perfectly clear throughout. After lying in this helpless condition for hours, he gradually recovers the lost motor power, and toward evening of the same day is able to get up, the power usually returning first in the arms, after which a complete return to the normal takes place. Speech is always unaffected; there is no difficulty in swallowing and never the slightest pain.

From this time until the onset of the next attack the patient regards himself, and his friends regard him, as a normal individual, capable of the same muscular exertion as his companions. He has always noticed that any overexertion tends to bring on an attack, which must, however, be preceded by a period of rest. In this way he has come to predict, with relative certainty, the occurrence of a "lameness," as he calls it. He usually pays the penalty for a particularly enjoyable day of activity by an enforced rest of many hours the following day, the onset of the paralysis coming some time during the night. He is able to abort a threatened attack temporarily during the day by increased muscular exertion, but it is merely a postponement of the inevitable paralysis, which follows with all the more certainty later. He is not able to attribute the attacks to dietary errors, or to anything whatever beyond the mere fact of exertion.

Physical Examination.—I first saw the patient April 8th, 1897, between attacks. He presented a picture of perfect health, and the examination, as made at that time, showed nothing which could be regarded as abnormal. His muscular system was particularly well developed; the legs somewhat out of proportion to the arms and body. His calves were noticeably large. Measurements made at the time were as follows: Chest under the nipples, 31½ inches unexpanded; 33½ expanded; biceps, midway between shoulder and elbow, right, 11½; left, 10¾ inches. Thigh, a little above patella, right, 20½; left, 20 inches; calf, maximum circumference, right, 16½; left, 16¼. No pathological significance was to be attached to these measurements.

June 18th, 1897. Saw the patient again at his home, which was some distance from Boston. The previous day he had ridden fourteen miles on a bicycle and otherwise exerted himself, but went to bed feeling well. He awoke at 6 the following morning, completely paralyzed, except for the slightest possible movements of the hands and feet. The face was unaffected. He remained in this helpless condition until about three in the afternoon, when a gradual improvement began, first in the arms. Owing to the distance at which he lived, I was unable to see him until about five in the afternoon, after improvement had begun. He was then in a state of partial paralysis. Movements were made with the greatest difficulty and weakness. The legs were almost completely beyond his control; the arms less so; neck muscles very weak. He could raise himself in bed only with the greatest difficulty. His efforts reminded me of a person in the last stages of a prostrating disease. His mind was perfectly clear, and he made no complaint of pain. It was a hot day, and he was sweating profusely. His face was somewhat flushed, but his temperature was normal. Pulse 88, and of fair quality. The heart[8] sounds were normal; the pupils were equal, of normal size and good reaction; swallowing was unaffected. He was totally unable to hold his head up, unsupported. All movements of the arms extremely weak; when he was supported in a sitting position he could not raise his arm above the horizontal. The legs were much weaker than the arms. Slight flexion at the knees was possible, and extremely slight flexion at the ankle. Flexion and extension of the toes possible, but very weak. Feet slightly adducted. Muscles all felt normal. Difficulty in coughing and sneezing.

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[8] In view of Oppenheim's work, a more careful examination of the heart is to be desired.

Reflexes: Knee-jerks lost; no plantar; cremaster very slight; abdominal and epigastric, active.

Electrical Examination; the faradic current alone available: Marked quantitative diminution of reaction from the facial nerve. Right arm: median, ulnar and musculo-spiral (?) nerves tested. Slight reaction from very strong currents. Same from Erb's point. Muscles: Deltoid, sterno-mastoid, trapezius, flexors and extensors of forearm, interossei of the hand, excited to contract only by very strong currents. Right leg: Very slight reaction from peroneal nerve; none from the posterior tibial nerve. Muscles: Vastus externus, quadriceps, interossei, slight. Left leg: Even less reaction obtained from the same muscles and nerves. The strength of the current used was sufficient to produce painful tetanic contraction in a normal person.

The result, therefore, of the partial electrical examination, which was alone possible, was enormous quantitative diminution to both indirect and direct stimulation. No qualitative change to faradism was noted. There was no disturbance of any form of sensation, and the patient took an intelligent interest in the examination throughout, answering all questions with perfect clearness.

At six in the evening, twelve hours after waking, he had eaten nothing but a few bananas, and had not had the slightest desire for drink. He had not passed urine for twenty hours, nor had an operation of the bowels for at least forty-eight hours. An hour later, at about 7 o'clock, he was again examined, and it was found that an increase in the weakness had again come on, an unusual occurrence. It was probably to be explained by the effort of muscular exertion, occasioned by the preceding somewhat prolonged examination.

Two days later, June 20th, patient wrote the following letter, which I reproduce in part: "I thought I would write and tell you how I came out of my immovability. When you left I had grown somewhat weaker than when you came, as you know, but that was only a warning of what was to come. In the night it came back full force. I was unable to sleep very long at a time. It is not exaggerating to say that I was truly miserable until it left me, about 3 or 4 o'clock Saturday morning. I was so weak and sore that I did not get up until about 11; the rest of the day I took life fairly easy, but was a little tired when I went to bed. This morning (forty-eight hours after first onset) when I woke up, I was a little lame, but after I got up it worked off, and I am feeling quite well now."

The fact of his being able to write in a steady hand shows the complete disappearance of the paralysis from the hand muscles.

Several months later, September 19th, 1897, his aunt, with whom he lives, wrote as follows: "I thought you might be interested to hear of J.'s condition since we were at your office. The following day, Wednesday, he was unable to dress until 3 P. M.; Thursday, not until 5 o'clock; Friday, at 10 o'clock, so went to school at recess. Saturday he was all right; to-day (Sunday), able to get out of bed at 3:30 P. M. He wished very much for me to send for you to-day, etc. Mrs. G.[the sister about to be described] was also in bed the greater part of the day yesterday, only able to sit up about an hour the latter part of the afternoon."

May 11th, 1898, the patient J. was again seen during a free interval. A careful examination resulted negatively. The attacks had diminished somewhat in frequency for the past few months. He is to-day feeling perfectly well.

Physical Examination: Muscular system apparently normal; no impairment of strength anywhere. Mechanical irritability of muscles, normal. Knee-jerks present and normal. Peripheral reflexes obtainable, and sufficiently active.

Sensation undisturbed; pupillary reactions normal; no enlargement of the spleen.

Heart perfectly normal as regards size and sounds. Pulse, 88, good quality.

Electrical Examination: Strong currents well borne. Considerable skin resistence. *Faradism:* Prompt and normal reaction from nerves and muscles of arms examined. Good reaction also from nerves and muscles of legs, with the possible exception of the peroneal group in the right leg. Inasmuch, however, as the indirect stimulation from the peroneal nerves produced reaction of the muscles supplied by it, we would not lay particular stress upon the poor response of the muscles to direct stimulation. *Galvanism:* Normal reactions, with the same exception above noted. Response quick; cathodal contraction much greater than anodal. No polar changes nor slightest indication of a reaction of degeneration. No exhaustion of muscular contraction through prolonged stimulation. In general, somewhat stronger currents were necessary than in case of the control used, but not more than could be explained by a heightened skin resistence.

Blood: Examination of the blood was kindly made for me by Dr. H. F. Hewes, of the Harvard Medical School. His report of a specimen taken between the attacks is as follows: "Probably no increase in white corpuscles. In a differential count of 200 white corpuscles, basophiles 51%; polymorphonuclear neutrophiles, 47%; polymorphonuclear eosinophiles, 2%; no myelocytes nor other pathological forms. Summary: Lymphocytosis, no evidence of anæmia. The large proportion of basophiles is characteristic of a reduced condition of nutrition. The count would be normal for ten years of age."

In a personal note Dr. Hewes writes: "The examination of the blood of Mr. S. shows practically no evidence of anæmia or of degenerative changes in the corpuscles. The lymphocytosis suggests lack of vigor in the metabolic processes, that is, if the patient is over 10 years of age. It is a condition similar to that found in the blood of rachitis of strumous children," etc.

An examination a few days later, also in a free interval, showed in a differential count of 200 white corpuscles: Basophiles, 57%; polymorphonuclear neutrophiles, 42%; polymorphonuclear eosinophiles, 1%; a few endoglobular poikilocytes and microcytes were seen. No malarial plasmodium. Very slight suggestion of loss of corpuscular substance in red corpuscles, evidently leucopænia plus lymphocytosis.

Of this specimen Dr. Hewes writes: "The relative increase in the younger forms of leucocytes is more marked than in the other specimen. The form is mostly that of the small lymphocyte. There is evidently a very small number of white corpuscles, perhaps four thousand per c.m. of blood. This also is a sign of lowered vigor."

Urine examination between attacks, also by Dr. Hewes; I append his report:

"May 19th, 1898, mixed twenty-four-hour specimen. Quantity of urine in twenty-four hours, 1,170 cc.; color, normal; specific gravity, 1,022; reaction, acid; amount of sediment, slight; urophæn, normal; chlorides, low; indoxyl, increased; sulphates, normal; urea, 1.85%; earthy phosphates, diminished; uric acid, normal; alkaline phosphates, diminished; albumin, absent; sugar, absent; bile pigments, absent; no acetone; total chlorine, 8.5 gms.; total urea in twenty-four hours, 21 gms.; sediment, centrifugal; few small, round epithelial cells; no casts found; a fresh specimen examined one week ago showed nothing abnormal.

"The urine in this case suggests a somewhat low condition of metabolism. The urea excretion is below normal for a man of this age and weight upon ordinary diet. It should approximate 30 grammes instead of 20. The twenty-four-hour amount of urine is low. The chlorine is low; normal phosphates are low; there is no special evidence of an excessive formation of uric acid in place of urea, nor of failure in carbohydrate metabolism; no sugar or acetone. Proteid metabolism is, as I have said, low. Fat metabolism can, of course, be judged by the stools only."

Owing to the distance at which he lives repeated examination of the patient during the attacks has not been possible. From the foregoing it will appear that the attack described was a severe one of a typical sort, and that the examination was made toward its close, or at least after the most extreme paralysis had begun to pass off. The signs observed were characteristic for that period, the most important being loss of knee-jerk, and great quantitative diminution in electrical excitability.

Case II.—Mrs. G., sister of previous patient. This patient I have been able to see but once, and then between attacks. She gives the following history:

She was at the time of the examination, September 14th, 1897, 24 years old; married; no children. When she was twelve years old, and much worried about the illness of her mother, on rising one morning she found it difficult to walk. The calf muscles seemed chiefly affected; she could not raise her heels readily. She went downstairs and fell from weakness, or stumbling, she does not know which. She made no mention of the feeling of weakness, and walked a considerable distance, experiencing the same difficulty. This continued during the day, and passed off the following day.

Two or three weeks later she had a second attack, which she attributed to taking medicine by mistake the evening before. In the morning she had the same trouble in walking. Toward night she could not walk upstairs, nor lift her feet. She was carried. The arms and face were unaffected. From this time on she had similar attacks, varying in frequency from one a week to one a month, and growing gradually more severe. A few months later she was completely helpless for the first time, the arms as well as the legs being affected. In the very worst attacks, of which she has had three or four, the face has been somewhat involved. She describes the disturbance as a drawing sensation in the lower part of the face, associated with a certain difficulty in opening the jaws, but not in closing them. She has never been unable to close her eyes. When her face was affected, she also had difficulty in breathing. She could not cough or sneeze. There was no disturbance with the special senses, nor with sensation generally.

Two years ago she took a "headache medicine" of unknown character in the evening. The following morning, at 9 o'clock, she suddenly lost consciousness while lying on the bed. She found herself about ten minutes later in a corner of the room. She did not bite her tongue, and had never had a similar attack, nor had she ever fainted in her life. After recovering consciousness from this attack, whose character is, of course, doubtful, she at once became generally weak, and forthwith went through the most severe paralytic seizure she has ever had. At 12 o'clock she was helpless. Her face was involved; she breathed only with the greatest difficulty, and had artificial respiration during the night. She was also given digitalis. The following day she was somewhat better, but still helpless. The next day, about forty-eight hours after the onset, she was able to get up, but remained very weak. She was not entirely strong for seven days. She had been married three weeks before this attack. During her wedding journey she had been travelling and walking much more than was her custom.

After this violent seizure she seemed to improve, and had milder attacks at longer intervals. This continued up to February, 1897. Then she had tonsillitis, and a serious attack, but less so than the one above described. This was followed by slight attacks up to a week ago (September, 1897), but for a month she has been feeling weak in the characteristic way. In September, 1897, a week before I saw her, she had another severe attack, coming on after a period of anxiety. She went to bed well, and woke up perfectly helpless, with both face and respiration affected; the attack lasted about twelve hours; four hours later she came to Boston. She states that her muscles always feel weak and sore after an attack.

Physically, she is an exceedingly strong and healthy looking woman. She has no disturbance in the bowel or menstrual functions, and, apart from her family infirmity, considers herself well. Owing to the relative infrequency of her attacks, she never lost work at school, and was the valedictorian of her class. She is a woman of considerable general culture.

Her pulse was 88; slightly irregular. She sleeps fairly, has no headaches, and is not of a nervous temperament. She has noticed no difference in the urinary secretion during and between the attacks.

This patient has not been seen since this visit in September, 1897. I have only learned from other members of the family that she is, on the whole, improving, but by no means free from the attacks. In average they are less severe than in Case I., although occasionally very much more so, even threatening a paralysis of respiration, as already described.

The two cases are evidently identical in their essential nature, though differing somewhat in detail. The examination has been in neither case, notably in the second, as complete as desirable, but amply sufficient to establish the diagnosis, and add, we hope, some points of interest to the literature already collected.

In consideration of the relative fewness of the cases reported, and extreme rarity and growing interest of conditions of this character, we offer the following abstract of cases hitherto appearing in literature, in the order of their publication:

Case I. Cavaré: Woman, 24 years old, several attacks of a general paralysis, lasting five to eight hours; quotidian type; relieved by quinine; no statement as to malaria.

Case II. Romberg: Case of paralysis of lower extremities, occurring in three attacks of the quotidian type. Disappeared on administration of quinine. Thought by Romberg to be due to intermittent fever (malaria).

Case III. Hartwig (quoted by Westphal): Male, 23 years old; healthy family; five years before had had tertian intermittent fever. Onset; "tired feeling" in legs; increased; spread to arms. On third day moved with great difficulty. Following night, paralyzed; legs, arms, body and movements of head. Facial muscles free. Speech, breathing, swallowing somewhat difficult. No incontinence. Sensibility unaffected; general condition good; no fever; some sweating; after twenty-four hours improvement in following order: neck, fingers, arms, body, legs. Similar attacks lasting about twenty-four hours. Attack later observed; muscles of respiration, except diaphragm, affected; coughing and sneezing impossible. Loss of reflexes. Electrical excitability almost abolished; at same time numbness, formication and cramp in paralyzed parts. Temperature normal. Paresis of extremities persisted and increased. Secondary contractures. Quinine merely temporary effect. Paresis increased by rest, diminished by movement. Patient lost fifteen pounds. (Later history not given.)

Case IV. Samuelsohn: Male, 38 years old; affection began at 18. At first, two attacks yearly; later one a week. Legs, body fully paralyzed; upper extremities only partially. Sensation undisturbed. In ten hours recovery. Quinine and other drugs unavailing. No electrical tests made, nor observations on the reflexes. Samuelsohn thought case of hysterical character.

Case V. Schachnowitsch: Father of patient similarly affected. Said to have died from increasing attacks. One brother epileptic. Patient, a man; attacks for twenty-five years; usually begin at night; also has abortive attacks indicated by subjective sensory disturbance and weakness; attacks could be checked by energetic movements; muscles tense.

Case VI. Gibney: Child of 7; after malarial attack, paralysis of four extremities. Sensory disturbances. Slow recovery, after months. Muscular atrophy. (A doubtful case.)

Case VII. Gibney: Child of 6; similar to preceding; slowly recovering paralysis; much constitutional disturbance; malarial. (A doubtful case.)

Case VIII. Westphal: Boy of 12; strong. Onset; weakness in legs with a pricking sensation. Great thirst and desire to urinate. Some minutes before he could pass urine. Three hours later could not stand. Slight movements of hips and arms possible; at midnight complete paralysis. Seen by Oppenheim at 4 A. M. Mind clear, cranial nerves free. Turning of head difficult. Coughing, sneezing impossible. Complete flaccid paralysis of arms and legs; sensibility normal. Plantar reflex lacking; cremaster and abdominal present; knee-jerk weak and not always obtainable. No dyspnœa.

Electrical examination: Faradism; strong, painful currents, very weak response from nerves. None from left peroneal nerve and its muscles. At 8 A. M. gradual return of reaction to G. and F., direct and indirect. Quick contractions when present at all.

Improvement: in evening could walk. No enlargement of spleen. Sweating. Temperature 37.7. Recovery on the following day, except for slight electrical changes in certain muscles. Similar attacks followed. Movement prevents onset of attack.

Etiology: No heredity. Scarlet fever followed some years later by unknown illness. Four weeks after exposure to draught sudden paralysis. Well between attacks which were at first four to six weeks apart, and lasted about twenty-four hours. Same patient studied again later by Oppenheim. Condition between unchanged. Attacks usually at night; awakes paralyzed, decreasing in intensity until following evening. Arms, legs, head paralyzed; face, tongue, larynx and eye muscles free. Thirst, heat, sweating. No excretion of urine. Intensity of paralysis not always the same. No pain; no disturbance of consciousness; no paræsthesia, except tickling in soles of feet. Swallowing practically unhindered, knee-jerks lost. Heart, slight temporary enlargement, and murmurs indicative of insufficiency. Heart normal between attacks. Electrical examination; results not uniform. In general, complete loss in certain muscle groups at height of attack. During attack often increased skin resistance. Later observations: slight atrophy of those muscles, with poorest electrical reaction between the attacks. Permanent weakness of legs.

Case IX. Fischl: Girl, 8 years old, good heredity. November, 1884, scarlatina, nephritis; complete recovery. Following May, lassitude, headache and backache, appetite poor. Somnolence, difficulty in rousing her from sleep. One morning could not stand or sit without support. Examination, mind clear; anxious expression; somewhat rapid pulse; temperature of head, body, arms normal; of legs and lower part of thighs ice cold, and remarkably pale; movements of head free. Cranial nerves free, movements of body good. Sitting up, or rising from prone position not possible. Arms normal when examined. Said to have been weak at first. Lower extremities completely paralyzed; flaccid. Sensation, diminished in affected areas. Abdominal reflexes present. Knee-jerks lost. Plantar reduced. No muscle excitability. No splenic enlargement; internal organs normal; urine and fæces normal, and normally passed. Attack lasted three hours. Sudden and complete recovery. Second attack, a few days later, also in the morning. Electrical examination: faradism; arms, normal; legs, no reaction to strong currents, excepting some muscular contraction in anterior crural distribution. Duration, five hours. Slower recovery. Normal reactions between attacks to galvanism. Third attack, less than an hour's duration. Same otherwise, as before. Fourth attack, length one half hour during day· arms weak, then legs. Recovery of arms first. Prodromata: headache, pain in back and legs, yawning, weakness. Later attack, twenty-four hours' duration, right foot only affected. Subsequent attacks irregular, characterized by drowsiness, one quarter to one half hour long, usually in afternoon. Child slept normally except for occasional stretching. Later, feeling of being tired. Subsequently became entirely normal (somewhat doubtful case).

Case X. Cousot: Mother and four children; no heredity otherwise. All strong, but smaller children of family of ten, affected. The mother often lost motor power at times twice weekly, through life.

Son, man now of 34; attacks began at 14, increased up to 20, then stationary. Strong in appearance; perfectly healthy on examination between attacks; attacks very frequent; usually began at night. First, feebleness of articulation; desire to walk; no sensory disturbance; usually arms last to lose power and first to regain it. Emotions increase tendency to attacks. Work and fatigue make them more violent. Duration, eight to ten hours; most helpless at third or fourth hour of attack. Muscles of face always spared. Deglutition and speech difficult. Attacks deferred by exercise. Intelligence unimpaired. Sweating during attacks. No urinary nor fæcal incontinence. Urine: no sugar, no albumin, much uric acid. Electrical reactions lost; galvanic response first appears on recovery.

The next Cousot case, not carefully observed; boy, onset at ninth year, similar to foregoing but more severe.

The other two cases, girls, onset in both about tenth year; precisely similar attacks.

Case XI. Griedenberg: Soldier; 22 years old; healthy; no heredity; similar attacks to foregoing; occasionally attacks limited to legs; cranial nerves free; certain muscles contracted.

Case XII. Pulawski: Man, 21 years old. Nothing significant in past history. Had had two previous short attacks. Present attack: went to bed well, at one A. M. could not move. No pain, sphincters unaffected. Examined in hospital; movements of arms not possible; slight movements of fingers. Same condition in legs. Could not sit up; lateral movements of head possible, not forward and backward. Muscles flaccid; respiration free, but superficial. Deep respirations and coughing impossible. Cranial nerves free. Sensation and muscle sense, normal. Knee-jerks lost. Cutaneous reflexes normal. Internal organs normal. Feeble faradic reaction. Hypnotism failed twice. No evidence of an antecedent intoxication. Urine, alkaline; specific gravity, 1017; no albumin, no sugar, abundant sediment of ammonium urate. Attack lasted one day; quinine given; following night much sweating, recovery.

Case XIII. Goldflam: Eleven cases in patient's family, on mother's side. No other neurotic heredity. Transmitted through both males and females. Onset usually between fifteen and twenty. Frequency varying from weekly to yearly attacks. Mother of patient had one attack in thirty-sixth year. Attacks generally more frequent in youth, but never cease altogether; in only one case was paralysis limited to legs.

Case reported in detail: Boy of 17. Previous history, typhoid fever, scarlet fever, with ear involvement, eczema; otherwise well. At thirteen paralysis of body, head, and extremities; duration three days. Saliva could not be swallowed. Following summer two equally severe attacks; later others, not so severe but oftener, lasting from twenty-four to forty-eight hours; then more severe, followed again by milder seizures. Attacks usually Friday, beginning at 6 or 7 P. M. First, weakness of legs, arms less affected, on well evenings much trouble with itching; none on evening of attack. No pain, chill nor fever. Constipation; complete paralysis, sparing head; thirst; sleepy during attack; consciousness otherwise undisturbed. Sense organs, speech, bladder, normal. Slight sweating during attack, more profuse toward its end. This, with acute itching for an hour ushers in end of attack. Return of movement in following order: Fingers, arms, body, legs, within a few hours. Capricious as to food, but no symptoms of intestinal disturbance. Drinks tea only on the evening before, and during attack. Physical examination: Flaccid paralysis, knee-jerks absent, plantar, abdominal and cremaster present. Sensation normal. No pain on pressure over nerves or muscles. No diminution in amount of urine during attack. No enlargement of spleen. Electrical examination: faradism; quantitative diminution in nerves of arms; loss in legs. No reaction from muscles in either arms or legs. Reaction of facial nerve normal. Mechanical muscular irritability lost. After attack, nerve and muscle irritability normal, excepting small muscles of thumb; slow contraction; An C=Ca C; R. D. chiefly in small muscles of hands.

Further study of above case made four years later: Itching less prominent than before. Prodromata now; feeling of coldness in legs, and then in arms. Occasional thirst; attacks usually at night. Abortive attacks, certain groups of muscles affected. Attacks occasionally aborted by active exercise, usually unavailing. In irregular attacks, relapses in stage of improvement. At times arythmic heart; systolic murmur at base. Slight accentuation of second tone; no enlargement; slow pulse; faint first tone in attacks.

Case XIV. Goldflam: Older brother of the foregoing; first attack at eighteen; attacks in general, similar; in one, dangerous asphyxia. Good muscular development. Electrical examination: direct and indirect irritability markedly diminished to both streams between attacks. Rapid fatigue of muscles. Slow contractions; polar changes. Ca C Te. obtained with weak currents. Faradic R. D. and partial R. D. In free intervals qualitative changes prominent; in attacks, quantitative. Loss in attack proportional to amount of paralysis. Mechanical irritability lost in complete attack. These changes in general found in both brothers. Goldflam mentions in a footnote to his article, to which reference has been made, two other cases in children, not yet adequately observed.

Case XV. Burr: Male of 30, no heredity whatever; no malaria. Began at 10. One attack about every four months since. Attacks last from one day to a week. Recovery gradual. Cranial nerves free. Slight involvement of neck muscles. Sensation, sphincters, mind normal. Attacks increasing somewhat in frequency. When seen by Burr, hemiparesis. Knee-jerk absent on left; present on right, but slight. Spleen not enlarged. No pain. Attacks always come at night; at times momentarily weak in the street, never at home. No electrical examination made.

Case XVI. Hirsch: Male of 26; no heredity, except mother had similar attacks, often of very quick recovery and with thirst during attacks. Patient previously well; first attack at 19th to 20th year; arms first heavy and stiff, then legs; obliged to go to bed. Paralysis not extreme. Well in twenty-four hours. One such attack yearly thereafter until 1892. Prodromata of heavy and tired feelings in legs for days before actual attack. In 1894 first decided paralytic seizure. Arms, legs, body, face, free. Duration twenty-four hours; sudden recovery; thirst. Physical examination at hospital; mind clear; intelligence and speech unaffected. Facial and other cranial nerves free. Neck muscles markedly affected. Breathing superficial. Paralyses inspiratory and expiratory muscles, excepting diaphragm. Lungs normal. Heart dullness enlarged; first sound at apex impure, slight accentuation of second pulmonic. Insufficiency of mitral valve. Pulse regular, 78; abdominal muscles movable. Spleen normal. Complete paralysis of arms and legs; deep reflexes lost, also superficial reflexes, excepting abdominal, which were weak. Idio-muscular irritability of paralyzed muscles weaker than normal. Sensibility normal; also muscle sense. Occasional sharp pains in left foot; no painful points. Bladder and rectal functions normal. No albumin, no sugar, no increased sweating; normal temperature, good appetite. No electrical examination made. Rapid recovery during night; entirely recovered following morning; heart practically normal. Attack twenty-four hours coming on, forty hours' duration. Cause of attack as given by patient, "drinking of tea or grog the night before."

Case XVII. Rich: 22 cases occurring in five generations, no individual case particularly described. The type is as follows: Family affection, no other neuroses or psychoses in the family history. Cause, exposure to cold. Paralysis of tonic character following and affecting various groups of muscles, preferably the face. On one occasion complete motor paralysis, excepting tongue, from sleeping in moist underclothing. Paralysis of tongue may be brought about by holding snow in the mouth, taste remaining uninvolved. Sensation in general unaffected, mind clear. Desire to urinate; sphincters normal; complete temporary recovery with considerable antecedent weakness. The affection has never skipped a generation. No mention made of electrical changes, nor of reflexes during, or between the attacks (doubtful cases, belong rather to the congenital myotonias).

Case XVIII. Bernhardt: Man; affection began in extreme youth; sudden attack of general weakness. Difficulty always in walking; tendency to stumble. Some muscles hypertrophic. Three years later general weakness; electrical reactions diminished; mind clear (doubtful case).

Case XIX. Bernhardt: Son of preceding case; almost daily attacks. Tendency often to cough, but not possible; cyanosis, threatened suffocation, sweating. Father similarly affected. Examination three years later: Attacks on waking, going to bed well. Immovable; sweating; no loss of consciousness. Speech at times slightly affected. Two years later unstable, but special senses normal. Slight quantitative diminution to electrical current. Knee-jerk present; walks with great difficulty. No difficulty in rising from seated position. Bladder and rectal functions normal. No albumin, no sugar. Attacks similar, but less frequent than formerly. Never come on when exercising, only after rest. Neck muscles affected, cranial nerves free. Attacks last about fifteen minutes (somewhat doubtful case).

Case XX. Goldflam (reported in 1897): Man of 22; attacks began in eighth year, and have increased in frequency. Attack usually begins in the evening with weakness of arms and general feeling of exhaustion. Sleep usually follows; on waking, often complete paralysis. Duration, twenty-four to forty-eight hours. Mind, speech, dedeglutition, sphincters, free. Recovery begins in evening. At times attacks sudden in onset. Attacks much more frequent in summer than in winter. Muscles well developed apparently, but strength diminished in free intervals; also changes in electrical excitability, including muscles of face. Examination during an attack showed usual conditions; toward end of attack, urine had traces of albumin.

Case XXI. Goldflam: Boy of 7½; brother of previous case. First attack in fifth year, examination incomplete; certain electrical changes in free interval demonstrated.

Case XXII. Goldflam: Man of 28; first attack in eleventh year. Infrequent attacks of from twenty-four to seventy-two hours' duration. Attacks at night. Extremities, body and neck muscles paralyzed. Coughing difficult, appetite poor, mind and sphincters unaffected. Marked electrical changes; slow contraction.

Case XXIII. Goldflam: Woman of 25, sister of previous case. Onset of attacks at twenty-four. Hands, arms, legs affected, in order named. Paræsthesia in fingers. At times complete paralysis. Intelligence, speech, sensibility, sphincters, unaffected. Loss of appetite; recovery first in arms, followed by head and legs. Diminution to complete loss of reflexes during attack, with marked quantitative diminution in faradic excitability. Pulse irregular, rapid; heart murmur; no enlargement of heart. Trace of albumin in urine during height of attack.

Cases XXIV and XXV. Taylor: Complete report already given.

(To be continued.)

219. SYNDROME SYRINGOMYELIQUE AVEC HEMIATROPHIE FACIALE ET TROUBLES OCULO-PUPILLAIRES (The Syndrome of Syringomyelia with Hemiatrophy of the Face and Oculo-Pupillary Disturbance). Queyrat et Chretein (La Presse Médicale, December 24th, 1897.

The patient had had in infancy a series of abscesses of the skin. He did not walk until three years of age, but otherwise appeared well, and displayed normal intelligence. At the age of eighteen years he had a very painful panaritis upon the index finger of the right hand, causing the loss of the terminal phalynx. Three years later he had a second panaritis upon the left thumb, much less painful, but causing the same mutilation. At this time there was some loss of power in the left arm and a succession of panaritides, and he was treated by various physicians for leprosy without any improvement. His condition in 1896 was as follows: There was kyphosis and slight scoliosis in the dorso-cervical region, mutilation of the thumb, index and middle fingers of the left and of the thumb and index finger of the right hand. Some very large cicatrices, representing the situations of the infantile abscesses were found upon the arms. The face was markedly asymmetrical, the left side being distinctly smaller than the right, but there were no motor disturbances other than those produced by the difference in size. The hair grew equally well on both sides. There was slight paresis, chiefly in the arms. The right hand was somewhat weaker than the left. Reactions of degeneration were present in the muscles of the hands. Sensibility was diminished in the left half of the face, the left shoulder and the left arm and hand, most markedly in the latter situation, and involved touch, pain and temperature. The tactile sensibility was preserved in the right forearm, but the pain and heat senses were lost. Both the pupils were myotic, reacting well to light, but poorly to accommodation. The visual field was normal; the patient, however, was color blind, not being able to perceive green. The case is interesting on account of the existence of facial atrophy and the possibility of leprosy, although a careful search for the microörganisms in an excised piece of skin was negative. SAILER.